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The pivotal sign of CANVAS

Petersen, J A ; Wichmann, W W ; Weber, K P

Abstract: A 75-year-old woman complained about insecure gait since age 55. Clinical examination revealed signs of cerebellar ataxia, bilateral vestibulopathy, and peripheral sensory impairment. Sensory nerve action potentials were absent. The visually enhanced vestibulo-ocular reflex (VVOR) was impaired (video on the Neurology(®) Web site at www.neurology.org, figure 1) and the diagnosis of cerebellar ataxia (figure 2) with neuropathy and bilateral vestibular areflexia syndrome (CANVAS) was made.(1) CANVAS is considered to be a recessive disorder with a mean age at onset of 60 years.(2) VVOR impairment is its characteristic clinical sign.(2) It can only be elicited if both smooth-pursuit eye movements and the vestibulo-ocular reflex are deficient. Normally, both are redundant at low head velocities.(2.)

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The pivotal sign of CANVAS

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J.A. Petersen recorded the video and wrote the manuscript.

W. Wichmann acquired the MRI and revised the manuscript.

K.P. Weber designed the study, acquired the eye movement data and revised the manuscript.

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J.A. Petersen reports no disclosure.

W. Wichmann reports no disclosure.

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A Patient Consent-to-Disclose Form has been obtained for the video recording.

Case summary

A 75-year-old woman complained about insecure gait since age 55. Clinical examination revealed signs of cerebellar ataxia, bilateral vestibulopathy and peripheral sensory impairment. Sensory nerve action potentials were absent. The visually enhanced vestibulo-ocular reflex (VVOR) was impaired (Video, Figure 1) and so, the diagnosis of cerebellar ataxia (Figure 2) with neuropathy and bilateral vestibular areflexia syndrome (CANVAS) was made (1). CANVAS is considered to be a recessive disorder with a mean age of onset at 60 years (2). VVOR impairment is its characteristic clinical sign (2). It can only be elicited if both smooth-pursuit eye movements and the vestibulo-ocular reflex are deficient. Normally, both are redundant at low head velocities (2).

References

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2. Szmulewicz DJ, Waterston JA, MacDougall HG, et al. Cerebellar ataxia, neuropathy, vestibular areflexia syndrome (CANVAS): a review of the clinical features and video-oculographic diagnosis. *Ann N Y Acad Sci*;1233:139-147.

Figure 1. Video-Oculography of impaired VVOR and pathologic head impulse test.

(A) Slow head rotations demonstrate an impaired VVOR with multiple catch-up saccades (arrowheads) instead of smooth compensatory eye movements. (B, C) The video head impulse test shows bilaterally reduced VOR gains with overt catch-up saccades (arrows) after head rotation to either side.

Figure 2. T2-weighted sagittal MRI illustrating the cerebellar atrophy in CANVAS.

Marked cerebellar atrophy involving the vermal lobules VI, VIIa, and VIIb. Note also the spinal atrophy.

Figure 1

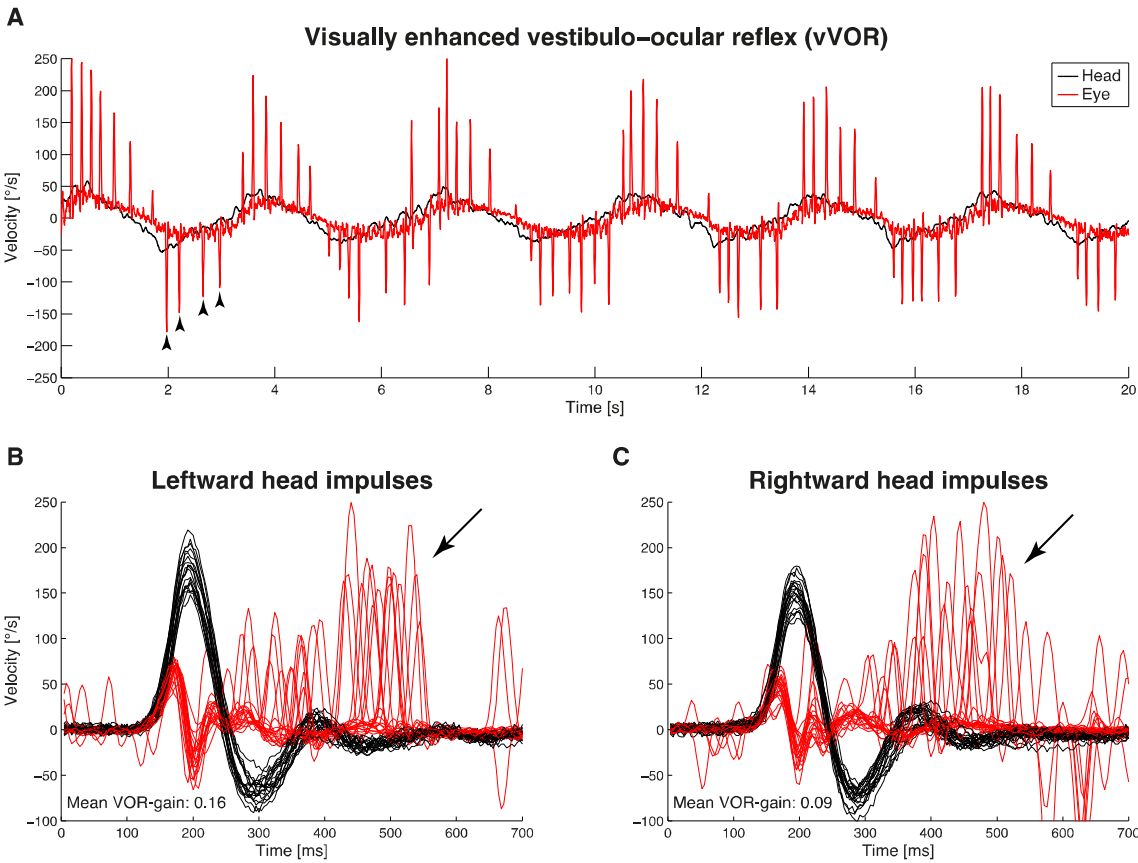


Figure 2

